One heart is not enough – conjoined twins: prenatal diagnosis and assessment of associated malformations. Case report

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Abstract

The incidence of multiple pregnancies has increased over the years due to the improvement of reproductive techniques, being usually a reason to rejoice for the parents, although, from a medical point of view, it is considered a high-risk pregnancy. One of the strangest and rare complications is that of conjoined twins, such as one case diagnosed, monitored and born at the Department of Obstetrics and Gynecology of the "Elias" University Emergency Hospital, Bucharest. The unfortunate outcome is marked by the various associated anomalies and by the complete and detailed examination of the pregnancy.

Keywords: *conjoined twins, parapagus dicephalus, twin monozygous pregnancy, surgical separation*

Rezumat

Incidența sarcinilor multiple a crescut de-a lungul timpului datorită îmbunătățirii tehnicilor de reproducere asistată și reprezintă de obicei un motiv de bucurie pentru părinți, cu toate că, din punct de vedere medical, este considerată o sarcină cu risc crescut obstetrical. Una dintre cele mai rare complicații ale acestui tip de sarcină este cea în care fetii sunt conjuncți, așa cum a fost cazul unei sarcini diagnosticate, monitorizate și născute la Clinica de obstetrică și ginecologie a Spitalului Universitar de Urgență „Elias" din București. Rezultatul nefavorabil, deși așteptat, a depins de anomaliile complexe și variate, precum și de monitorizarea detaliată și multidisciplinară a cazului.

Cuvinte-cheie: *gemeni conjuncți, parapagus dicephalus, sarcină monozigotică, separare chirurgicală*

Submission date: 20.02.2021
Acceptance date: 25.02.2021

O singură inimă nu e de ajuns – gemeni conjuncți: diagnosticul prenatal și evaluarea malformațiilor asociate. Prezentare de caz

Suggested citation for this article: Munteanu AE, Constantin ME, Vlădăreanu S, Tecuci A, Pop-Began C, Vlădăreanu R. One heart is not enough – conjoined twins: prenatal diagnosis and assessment of associated malformations. Case report. Ginecologia.ro. 2021;31(1):12-15.

A 36-year-old female patient, Caucasian, presented at 13 weeks of amenorrhea at the Department of Maternal Fetal Medicine of the "Elias" University Emergency Hospital, Bucharest, for first-trimester anomalies screening. Both she and her family had unremarkable medical histories. Her personal obstetrical history revealed two vaginal live births without any complications, with normally developed fetuses. The pregnancy was obtained by spontaneous conception, and there was no history of twin pregnancies in the family. An ultrasonographic diagnosis was established, using both transabdominal and transvaginal probe of the ultrasound machine Samsung WS80A: monochorionic monoamniotic twin pregnancy, *parapagus dichephalus*, 13 weeks and 2 days of amenorrhea, with one fetus with two heads (Figure 1), two spines fused at the sacral region (Figures 2-3), one heart, two superior limbs and two inferior limbs.

The detailed transabdominal scan, although limited by the early gestational age, described one normal head and the other with cystic hygroma, two necks and joined thorax, abdomen and pelvis. The fetus was in a fixed position with reduced movement. At the level of the fetal thorax and abdomen, there were described a compound heart placed centrally, one stomach on each side of the abdomen (Figure 3 a, b), normal entrance of the umbilical cord, normal abdominal wall, and two umbilical arteries surrounding the bladder. One of the stomachs was apparently normally placed while the other was situated on the opposite side, lateral and closer to the spine, excluding the presence of a double spleen. The other components of the gastrointestinal system could not be properly examined due to the early gestational age.

The heart was deviated to the center and the color Doppler evaluation of the four-chamber view was abnormal, with normal anterograde flow on the left side and diminished filling on the right side, the outflow tracts appeared to be doubled, with one big vessel outgoing from the left side, and one smaller, with retrograde flow leaving the right side, while the *ductus venosus* was present with reversed A wave (Figure 4 a, b, c).

The patient was further recommended medical and psychological support concerning the prognosis of the pregnancy and the high incidence of the fetal morbidity and mortality.

The management options for the pregnancy were discussed, which included expectant management with genetic testing or the termination of pregnancy, and the parents chose to continue the pregnancy with palliative care. Further on, for better monitoring of both mother and fetus, a multidisciplinary team was established, consisting of an obstetrician, a pediatric surgeon and a neonatologist. The patient was regularly examined, and the laboratory tests were within normal range. The second-trimester anomaly scan confirmed the diagnosis and brought additional information. A detailed examination was performed, and the following were observed: the fetal brain and face had normal structural development, and one of the fetal profiles

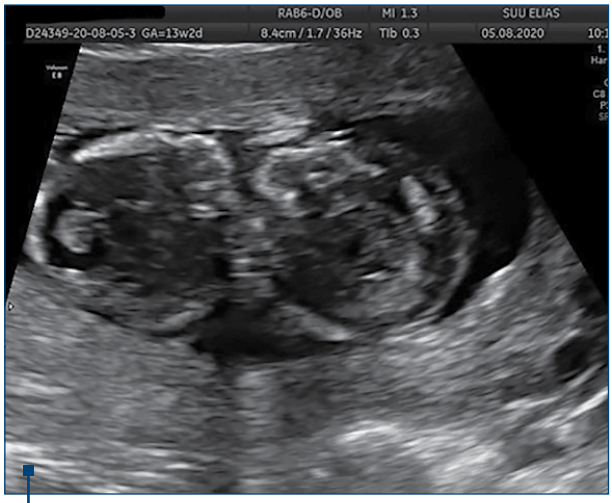

Figure 1. *Axial section of the separeted fetal heads*

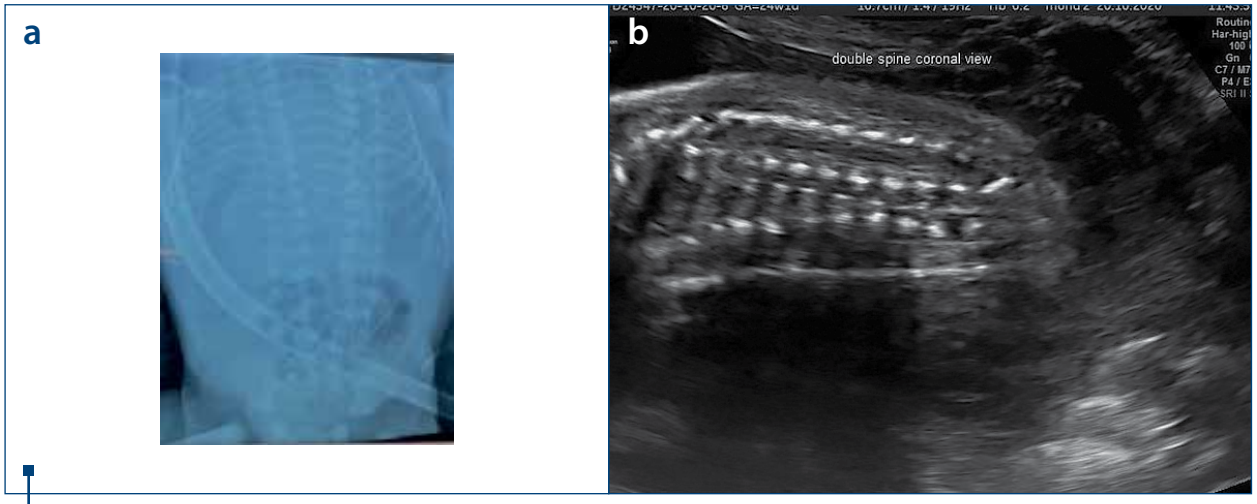

Figure 2. a) *X-ray of the newborn's spines that fuses in the sacral region;* ***b)*** *sonographic coronal view of the spine*

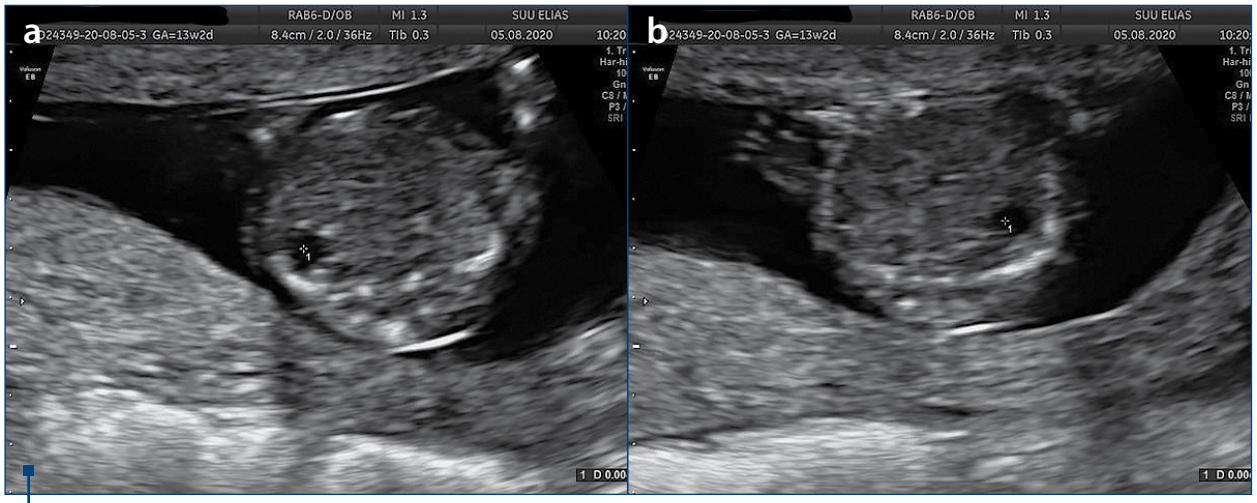

Figure 3. a) *Axial cross-section of the fetal abdomen showing a right posteriorly placed stomach;* ***b)*** *the same axial cross-section showing a normally placed stomach*

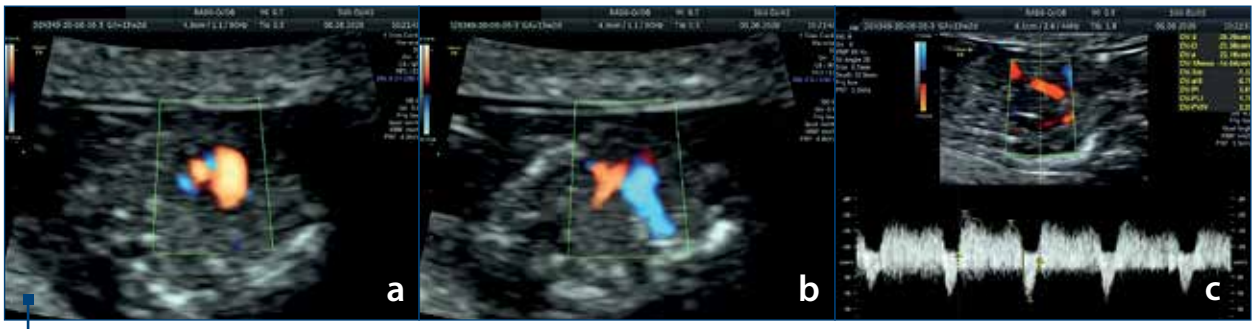

Figure 4. a) *First-trimester color Doppler examination of the four-chamber heart section;* ***b)*** *color Doppler of the outflows;* ***c)*** *sagital examination of ductus venosus showing reversed A wave*

was flattened due to fixed and extended cervical position (Figure 5 a, b).

The thorax was large, and the four-chamber view was abnormal. The heart axis was deviated, the left ventricle was enlarged, the outflow tracts were transposed and originated from the left ventricle. The aorta was anterior and large, the pulmonary artery was smaller and posterior. The right ventricle was smaller, with a thickened tricuspid valve, and another outflow tract with reversed flow, resembling a smaller aorta, connected to it (Figure 6).

It appeared to be one big kidney on the right side and a smaller pelvic left kidney. The liver was enlarged and occupied almost the entire abdomen. There were no obvious signs of intestinal obstruction, the gender was female, and the limbs appeared normal. Fetal growth could not be properly estimated, the amniotic fluid was normal, but the fetal movements were exceedingly reduced, with a fixed breech position and extended neck. At 37 weeks of gestation, the patient went into labor, and a caesarean section was performed. The delivered fetus weighed 3300 grams, with length of 45 cm, fetal heart rate of 100 beats per minute, and unfortunately died in the next hour.

Discussion

Conjoined twins, a rare complication of a monoamniotic monochorionic pregnancy, are usually defined by the two individuals that share one or more organs and parts of their anatomy. The real incidence is unknown due to miscarriages, and the reported range varies between 1:50.000 and 1:100.000 live births. There seems to be a significant predominance of the female gender in these cases[1]. The highest incidence is encountered in countries where the termination of pregnancy is illegal.

Etiopathogenesis of conjoined twinning

Because it's a rare event, the exact pathogenesis of this condition remains unknown, and still fascinates the scientific world, with theories regarding the genesis of conjoined twins. Up to now, there are two major theories: fusion and fission. The fusion hypothesis was the first to be proposed and it assumes an incomplete division of a single zygote, the formation of two cellular masses within the blastocyst at the end of the second week post-conception. Therefore, a sole embrionary structure is formed, but with a single yolk sac. The DNA analysis always describes a homozygous karyotype[2,3]. The fission theory assumes the incomplete division of the internal mass of the blastocyst followed by fusion on the 14-15th

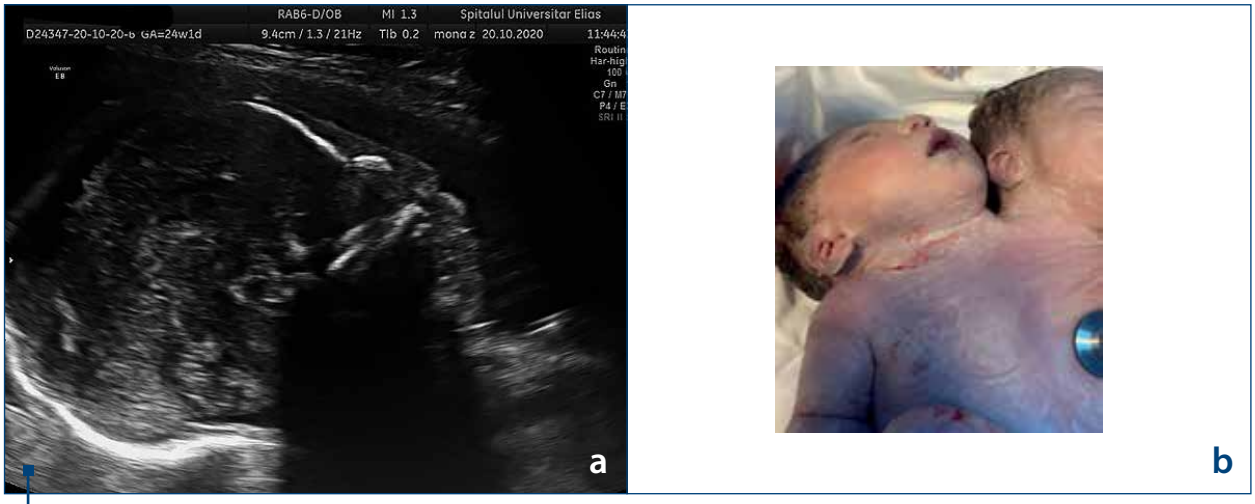

Figure 5. a) *Ultrasonographic sagital view of the fetal profile;* ***b)*** *parapagus dicephalus conjoined twins profile*

day post-conception[2]. Currently, the subject of conjoined twins is divided between symmetrical and asymmetrical conjunction. The latter consists of gross underdevelopment of one of the twin members, commonly known as "parasites" or "heteropagi"[4]. The most used classification divides symmetric conjoined twins into four general conjunction groups: ventral, lateral, caudal, and dorsal conjunction. The case presented consists in lateral conjunction which can involve the head, face, neck, thorax and abdomen, therefore it implies a greater risk for the pregnancy due to the organs that are shared. In this category, there are two subcategories: *parapagus dicephalus* (two heads) and *parapagus diprosopus*, which implies one head with two lateral oriented faces.

Diagnostic challenges

The clinical diagnosis of conjoined twins is close to impossible. The use of auscultation or palpation can only suggest separate twins, and in areas where the access to basic technology is limited, the diagnosis can only be confirmed at birth.

With the advances in ultrasonography, the early depiction and the proper prenatal evaluation of this condition is needed for better counseling the parents in order to offer the possibility of termination of pregnancy and, if the pregnancy continues, to establish a plan of delivery in a specialized center, improving the chances of survival and accomplishing the medical aim to surgically separate the twins. The earliest time of diagnosis is placed in the first trimester, and the criteria include, firstly, the absence of separating amniotic membrane, lack of movements, at different times of examination, of the body, fetal head or body parts, unusual proximity of heads or spines, bifid appearance of the fetal pole and, secondly, complex anomalies, compound heart, joined pelvis, incomplete limbs and multiple umbilical arteries[6]. In cases where the pregnancy continues, given the vast anomalies encountered, in order to detail the exact level of conjunction, anatomy, vascularization and potential for postnatal surgical separation, studies have shown that the use of 3- or 4-dimensional ultrasound, magnetic resonance imaging or computed tomography is indispensable.

Considering the vast union between the twins in lateral conjunction, the chance for separation is close to null, therefore the expected survival in the neonatal period is extremely low[5]. From the many types of conjoined twins, the lateral fused twins associate frequently the most complex anomalies due to disturbed cross-signaling between tissues, and past studies described at autopsy that when the heart is compound, there can be transposition or reversal of great arteries, right aortic arch and defects of laterality, such as absent spleen or *situs inversus*. The prognosis in usually poor, only 18% of all conjoined infants survive, approximately 35% of live births die within the first 24 hours, and only 18% of all conjoined twins survive longer than 24 hours[7]. In terms of delivery, although there are several reports of vaginal delivery, mostly for premature, small or nonviable fetuses, a planned caesarean section is preferred to avoid psychological and physical maternal trauma and to ensure the best chances of survival.

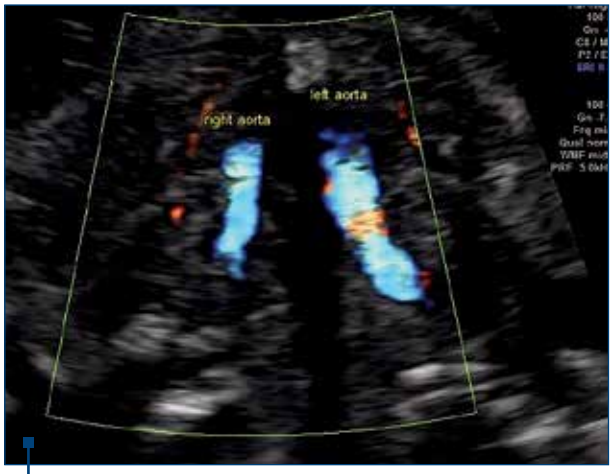

Figure 6. *Color Doppler of the outflow tracts*

Prognosis and postnatal management

The postnatal management of conjoined twins is nowadays surrounded by ethical dilemmas regarding the surgical approach. In the case of lateral conjunction, the management usually consists in palliative care because dicephalus twins are generally stillborn or die in a few hours after birth due to complex cardiac and pulmonary anomalies. Immediate surgical maneuvers are appropriate when removing a stillborn or critically ill twin's connection to save the healthier twin. In stable situations, the surgery, if wanted and needed, can be postponed until later infancy.

Conclusions

Conjoined twinning is a rare complication of monoamniotic monochorionic pregnancy. The vast variety of conjunction leads to different approaches regarding the prenatal and postnatal management. The prognosis is poor for lateral conjunction due to complex anomalies of vital organs, and the earliest diagnosis is necessary to ensure the parents of their options further on. In case the pregnancy continues, other means of evaluating the pregnancy are required and a multidisciplinary team must be organized. ■

Conflict of interests: The authors declare no conflict of interests.

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